



Romano-Ward syndrome

Romano-Ward syndrome is a condition that causes a disruption of the heart's normal rhythm (arrhythmia). This disorder is a form of long QT syndrome, which is a heart condition that causes the heart (cardiac) muscle to take longer than usual to recharge between beats. The irregular heartbeats can lead to fainting (syncope) or cardiac arrest and sudden death.

Frequency

Romano-Ward syndrome is the most common form of inherited long QT syndrome, affecting an estimated 1 in 7,000 people worldwide. The disorder may actually be more common than this estimate, however, because some people never experience any symptoms associated with arrhythmia and therefore may not have been diagnosed.

Genetic Changes

Mutations in the *KCNE1*, *KCNE2*, *KCNH2*, *KCNQ1*, and *SCN5A* genes cause Romano-Ward syndrome. These genes provide instructions for making proteins that act as channels across the cell membrane. These channels transport positively charged atoms (ions), such as potassium and sodium, into and out of cells. In cardiac muscle, ion channels play critical roles in maintaining the heart's normal rhythm. Mutations in any of these genes alter the structure or function of these channels, which changes the flow of ions between cells. A disruption in ion transport alters the way the heart beats, leading to the abnormal heart rhythm characteristic of Romano-Ward syndrome.

Unlike most genes related to Romano-Ward syndrome, the *ANK2* gene does not provide instructions for making an ion channel. The ANK2 protein, ankyrin-2, ensures that certain other proteins (particularly ion channels) are inserted into the cell membrane appropriately. A mutation in the *ANK2* gene likely alters the flow of ions between cells in the heart, which disrupts the heart's normal rhythm. *ANK2* mutations can cause a variety of heart problems, including the irregular heartbeat often found in Romano-Ward syndrome. It is unclear whether mutations in the *ANK2* gene cause Romano-Ward syndrome or lead to another heart condition with some of the same signs and symptoms.

Inheritance Pattern

This condition is typically inherited in an autosomal dominant pattern, which means one copy of the altered gene in each cell is sufficient to cause the disorder. In most cases, an affected person inherits the mutation from one affected parent. A small percentage

of cases result from new mutations in one of the genes described above. These cases occur in people with no history of Romano-Ward syndrome in their family.

Other Names for This Condition

- RWS
- Ward-Romano Syndrome
- WRS

Diagnosis & Management

Genetic Testing

- Genetic Testing Registry: Long QT syndrome 1
<https://www.ncbi.nlm.nih.gov/gtr/conditions/C0035828/>
- Genetic Testing Registry: Romano-Ward syndrome
<https://www.ncbi.nlm.nih.gov/gtr/conditions/CN119492/>

Other Diagnosis and Management Resources

- GeneReview: Long QT Syndrome
<https://www.ncbi.nlm.nih.gov/books/NBK1129>
- MedlinePlus Encyclopedia: Arrhythmias
<https://medlineplus.gov/ency/article/001101.htm>

General Information from MedlinePlus

- Diagnostic Tests
<https://medlineplus.gov/diagnostictests.html>
- Drug Therapy
<https://medlineplus.gov/drugtherapy.html>
- Genetic Counseling
<https://medlineplus.gov/geneticcounseling.html>
- Palliative Care
<https://medlineplus.gov/palliativecare.html>
- Surgery and Rehabilitation
<https://medlineplus.gov/surgeryandrehabilitation.html>

Additional Information & Resources

MedlinePlus

- Encyclopedia: Arrhythmias
<https://medlineplus.gov/ency/article/001101.htm>
- Health Topic: Arrhythmia
<https://medlineplus.gov/arrhythmia.html>
- Health Topic: Congenital Heart Defects
<https://medlineplus.gov/congenitalheartdefects.html>

Genetic and Rare Diseases Information Center

- Long QT syndrome 1
<https://rarediseases.info.nih.gov/diseases/3284/long-qt-syndrome-1>

Additional NIH Resources

- National Heart, Lung, and Blood Institute
<https://www.nhlbi.nih.gov/health/health-topics/topics/qt/>

Educational Resources

- American Heart Association
http://www.heart.org/HEARTORG/Conditions/Arrhythmia/AboutArrhythmia/Types-of-Arrhythmia-in-Children_UCM_302023_Article.jsp
- Centre for Genetics Education (Australia)
<http://www.genetics.edu.au/Publications-and-Resources/Genetics-Fact-Sheets/FS58PRIMARYARRYTHMOGENICDISORDERS.pdf>
- Cleveland Clinic
<http://my.clevelandclinic.org/health/articles/long-qt-syndrome>
- Disease InfoSearch: Romano-Ward syndrome
<http://www.diseaseinfosearch.org/Romano-Ward+syndrome/9638>
- KidsHealth from the Nemours Foundation
<http://kidshealth.org/en/teens/arrhythmias.html>
- MalaCards: scn5a-related romano ward syndrome
http://www.malacards.org/card/scn5a_related_romano_ward_syndrome
- Merck Manual Consumer Version: Long QT Syndrome
<http://www.merckmanuals.com/home/heart-and-blood-vessel-disorders/abnormal-heart-rhythms/long-qt-syndrome-and-torsades-de-pointes-ventricular-tachycardia>

- My46 Trait Profile
<https://www.my46.org/trait-document?trait=Long%20QT%20syndrome&type=profile>
- Orphanet: Familial long QT syndrome
http://www.orpha.net/consor/cgi-bin/OC_Exp.php?Lng=EN&Expert=768

Patient Support and Advocacy Resources

- National Organization for Rare Disorders
<https://rarediseases.org/rare-diseases/romano-ward-syndrome/>
- Resource List from the University of Kansas Medical Center
<http://www.kumc.edu/gec/support/conghart.html>
- Sudden Arrhythmia Death Syndromes (SADS) Foundation: Long QT Syndrome
<http://www.sads.org/What-is-SADS/Long-QT-Syndrome#.Vds7xZdGdD8>

GeneReviews

- Long QT Syndrome
<https://www.ncbi.nlm.nih.gov/books/NBK1129>

ClinicalTrials.gov

- ClinicalTrials.gov
<https://clinicaltrials.gov/ct2/results?cond=%22Romano-Ward+syndrome%22+OR+%22Long+QT+Syndrome%22>

Scientific Articles on PubMed

- PubMed
<https://www.ncbi.nlm.nih.gov/pubmed?term=%28Romano-Ward+syndrome%5BTIAB%5D%29+AND+english%5Bla%5D+AND+human%5Bmh%5D+AND+%22last+2160+days%22%5Bdp%5D>

OMIM

- LONG QT SYNDROME 1
<http://omim.org/entry/192500>

Sources for This Summary

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